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Case report/Kazuistyka

Pelvico-calyceal system rupture due to staghorn calculus with urinoma formation in a boy with neurofibromatosis type 1 and quadriplegia

Przerwanie układu kielichowo-miedniczkowego nerki na podłożu kamicy odlewowej z wytworzeniem urinoma u chłopca z nerwiakowłókniakowatością typu 1 i tetraplegią

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ABSTRACT

Nephrolithiasis is a rare condition in children. The urinary tract rupture related to stones formation or migration is atypical in children, but creates serious consequences. We present a case of a 17-year-old quadriplegic patient with neurofibromatosis type 1 and urinoma due to the rupture of calyceal fornices in the course of nephrolithiasis. The boy was admitted with symptoms of severe pneumonia complicated with sepsis and prerenal acute kidney injury. Abdominal ultrasound revealed stone casts in both renal pelvises. Antibiotics, fluid therapy and diuretics were used to improve patient's condition. On the 28th day gross hematuria was observed. The patient's condition was stable, without signs of pain or discomfort. Abdomen ultrasound showed heteroechogenic structure (125 mm × 100 mm × 100 mm) localized between the lower surface of the liver and the right kidney. Contrast CT scan confirmed urinoma under the right kidney capsula. Because of the high risk of its rupture, decision of invasive evacuation of perirenal fluid was made. Using the percutaneous catheter 700 ml of bloody fluid was drained. After 10 days catheter was removed without recurrence of urinoma. Concluding, in children with prolonged immobilization this condition should be taken into

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• *urinoma*
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consideration in differential diagnosis, also special attention should be paid for accompanying scarce symptoms.

Introduction

Nephrolithiasis in children is much less common than in adults. Alken et al. reported that pediatric stones account for only 1–5% of all urinary stones in the German population [1]. Up to 76% of pediatric patients with the diagnosis of kidney stone disease present metabolic abnormalities, most often hypercalciuria [2]. About 90–95% of kidney stones in children consist of calcium [3]. A specific condition related to high risk of urinary stones formation is a long-term immobilization due to severe neurological disorders.

Significant long-term consequences of nephrolithiasis include recurrent stone formation, urinary tract infections, progression of chronic renal dysfunction and finally the rupture of the urinary tract, most commonly ureters, with urine or blood leakage [4]. We report a case of a quadriplegic patient due to neurofibromatosis type 1 complications (brainstem tumor) with the kidney calyceal rupture in the course of nephrolithiasis, successfully treated with invasive procedures.

Retrospective analysis of medical records in a 17-year-old patient, including results of laboratory test, sonography, abdominal X-ray and computed tomography imaging was performed.

Case report

We present the medical history of a 17-year-old cachectic boy without logical verbal contact, with quadriplegia, epilepsy, and acquired hydrocephalus developed from the age of 13 as the complication of brain stem tumor in the course of neurofibromatosis type 1. He was admitted to the Pediatric Nephrology Department in severe general condition with the symptoms of sepsis, severe prerenal insufficiency and pneumonia. On laboratory examination, WBC was $30 \times 10^9\,l^{-1}$, C-reactive protein (CRP) level – 336.0 mg/l [normal range 0.0–5.0 mg/l], serum creatinine concentration – 353 μmol/l (which corresponded to eGFR value calculated according to Schwartz formula of 17.0 ml/min), serum urea level – 19.4 mmol/l, serum uric acid level – 540 μmol/l, and serum total proteins – 55 g/l. In the abdominal ultrasound stone casts in both kidney pelvises were found. Intravenous antibiotics and conservative symptomatic treatment were applied to achieve the improvement in patient's condition (blood test performed on 7th day: WBC – $23 \times 10^9\,l^{-1}$, CRP – 43.8 mg/l, serum creatinine – 111 μmol/l, and serum urea – 9.5 μmol/l).

At the 15th day of hospitalization patient presented anxiety, seemed to feel pain and significant discomfort in the abdomen. The ultrasound examination was comparable to the previous one. The abdomen X-ray revealed large amount of constipated stool in the bowel that confirmed the presence of stone casts in both kidneys, as well as showed the separated stone localized in the right kidney pelvic–ureteral junction and some small concrements at the projection of urinary bladder. There was no significant dilatation of pelvis and calyces (Fig. 1). Constipated stool was removed manually and then enema and laxatives simultaneously with analgesics and spasmolytics were given, leading to improvement of the symptoms.

At the 28th day of the hospitalization the episode of gross hematuria was observed. The patient's condition was stable; he did not show any symptoms of pain or other discomfort. Repeated abdomen ultrasound examination revealed oval, heteroechogenic structure, with dimensions of 125 mm × 100 mm × 100 mm, localized on the right abdominal flank, between the lower surface of the liver and right kidney. The presence of perirenal hematoma in retroperitoneal space has been suspected. In CT scan the collection of fluid with 11–58 Hounsfield units density under the right

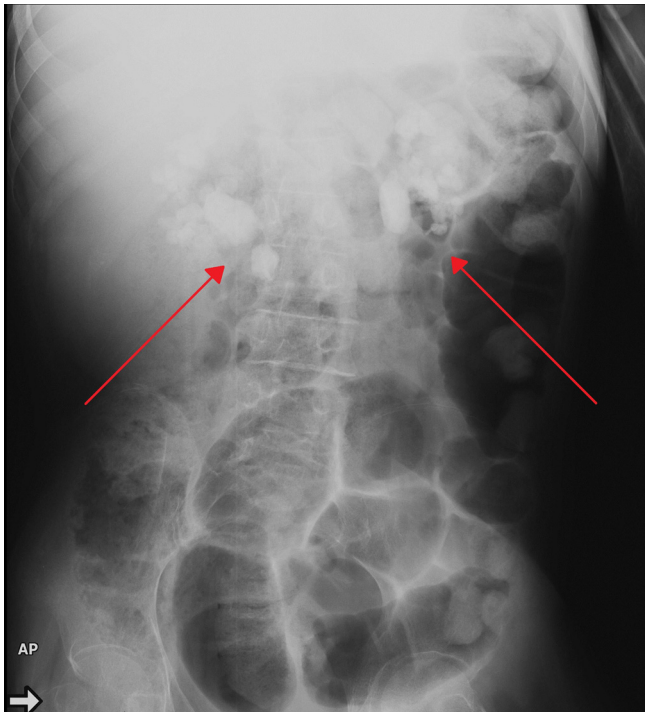

Fig. 1 – Abdominal X-ray. The presence of staghorn calculi in both kidneys and separated stones localized in the kidney pelvic–ureteral junction (arrows)
Ryc. 1 – Zdjęcie RTG jamy brzusznej. Kamica odlewowa obu nerek. Oddzielony złóg zlokalizowany w połączeniu miedniczkowo-moczowodowym (strzałki)

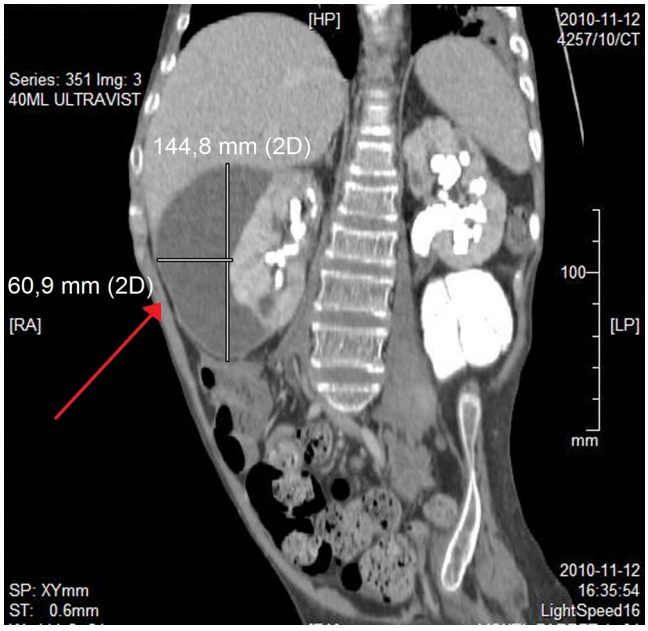

Fig. 2 – CT-scan of the abdomen. A large amount of fluid of 11–58 Hounsfield units density, under the right renal capsula (arrow)

Ryc. 2 – Badanie TK jamy brzusznej. Przestrzeń płynowa o gęstości 11–58 jednostek Hounsfielda pod torebką nerki prawej (strzałka)

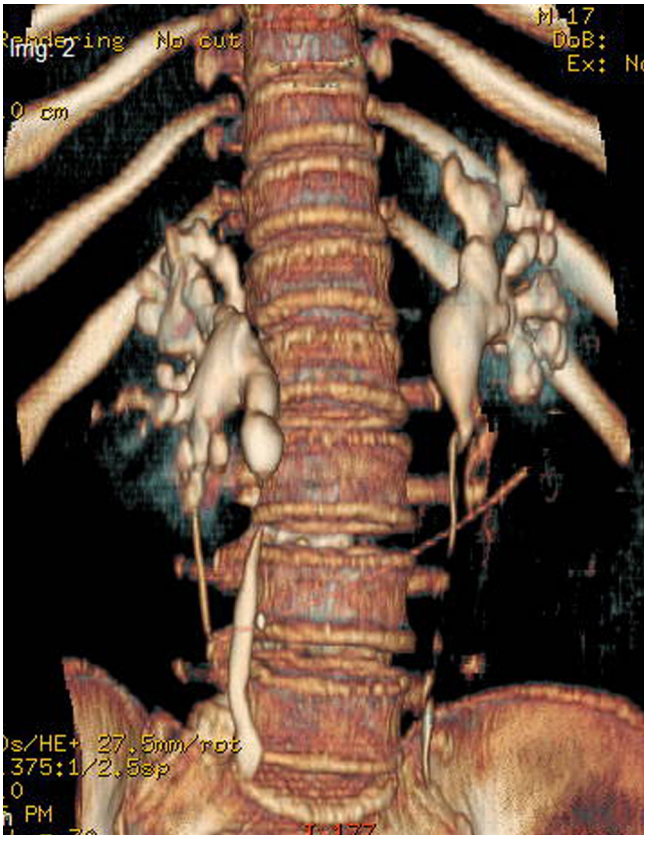

Fig. 3 – Abdominal CT. Reconstruction of staghorn calculi picture in both kidneys

Ryc. 3 – Badanie TK jamy brzusznej. Rekonstrukcja kamienia odlewowego w obu nerkach

renal capsule has been described (Fig. 2). In the arterial phase of contrast-enhanced CT examination there was no extravasation of contrast, and in delayed imaging the leakage of contrasted urine to the space limited by the right kidney capsula was noticed.

On the next day the small calcium oxalate-monohydrate stone was found in the urine container. Ultrasound examinations performed on consecutive days suggested progressive increase in diameter of the fluid structure up to 162 mm × 71 mm. Due to high risk of urinoma rupture, the decision of the surgical evacuation of the undercapsular fluid was made, despite the patient's stable condition and lack of any complaints. The percutaneous catheter was inserted on the 39th day, resulting in drainage of 700 ml of bloody fluid. During the following days the volume of the evacuated fluid was gradually reduced. Finally, at the 48th day of hospitalization the catheter was removed with no recurrence of urinoma and the patient was discharged from hospital.

Discussion

The urinary collecting system disruptions are usually caused by renal injury, pelvic mass, posterior urethral valves, or different bladder outlet obstruction, pregnancy, retroperitoneal fibrosis and transmitted back pressure due to obstruction of the urinary system by a ureteral stone [5–7]. It is also the result of iatrogenic injury, most often during extracorporeal shock wave lithotripsy (ESWL) [4]. According to Friedenberg et al. urinoma occurs if four risk factors coexist: preserved renal function, chronic partial distal obstruction which primarily interferes with high volume flow, renal calyces or fornices capable of extravasation during increased pelvic pressure and renal hilus that allows urine to extravasate outside of the kidney [8]. In our patient severe bilateral nephrolithiasis was present with staghorn stones in pelvises and multiple fine concrements (Fig. 3). The intravenous fluid therapy and diuretics used in the treatment of prerenal AKI, in the presence of the stone partially closing the outlet from the right kidney pelvis, could lead to increased pressure in the pelvico–calyceal system. However, the stone casts might have weakend the place of least resistance – the calyceal fornix, leading to its rupture and urinoma formation.

Several additional risk factors of urine stone formation due to secondary hypercalciuria could be found in our patient. The calcium excretion with urine examined during hospitalization remained within the normal range. However we cannot exclude former hypercalciuria. First of all he suffered from the progressive motor dysfunction due to brain stem tumor leading to tetraplegia at the age of 13 years. The immobilization can lead to increased demineralization of the skeleton. Such observations were documented in patients with traumatic spinal cord injuries, among whom the renal diseases were historically the leading cause

of death. The incidence of renal calculi in this group of individuals is assessed to be at 20%. The risk of urinary stone disease is especially high during the first 6 months after immobilization, when the bone mass resorption is the highest [9]. The other risk factor of hypercalciuria in the past history, present in our patient, is chronic treatment with glucocorticosteroids as the management of intracranial overpressure. Glucocorticoids increase bone resorption and sustain marked hypercalciuria leading to stone formation [10]. The next risk factor of the nephrolithiasis which could be observed in our patient might have been low fluid intake associated with inadequate nutrition. Despite the feeding by nasogastric tube, the patient was cachectic and his total proteins level in serum was below the normal limit. Therefore we can confirm that his nutrition was inappropriate for his demand. In children with neurological disorders, especially in patients with swallowing problems, severe caloric-protein malnutrition could often be seen [11, 12]. The problem is less common in patients fed by nasogastric tube or percutaneus endoscopic gastrostomy (PEG), however lack of appetite and thirst and the absence of self-feeding between main meals contribute to inadequate calories intake. Neurofibromatosis type 1 could be associated with some bone abnormalities as well as congenital kidney defects (horseshoe kidney, renal artery stenosis) [13–15]. However it seems that the disease per se is not a risk factor of nephrolithiasis. To the best of our knowledge, there is only one report of the association of neurofibromatosis type 1 with nephrolithiasis published so far [10].

The diagnostic problem we faced in our patient was the confounding clinical course of the presented complication. Patients with urinoma frequently present with clinical symptoms such as flank pain and haematuria; however urine leakage may be also clinically occult or from the other side leads to acute abdomen symptoms [4]. Our patient presented anxiety, some discomfort and abdominal pain 13 days before the haematuria occurred and urinoma has been found on ultrasound. The complaints seemed to be connected with chronic constipation and diminished after stool evacuation. We could not exclude that partial closing of the outlet from the right kidney pelvis was also a cause of pain and discomfort at this time. The gross hematuria which occurred on the day 28th of hospitalization could be the result of stone downward dislocation with the simultaneous injury of the urinary collecting system wall. However at this time no anxiety or discomfort was noted. We are not able to differentiate if sparse and confounding signs and symptoms of urine leakage and urinoma formation could be explained by the patient's neurological condition or just the clinically occult course of chronic disease.

Conclusions

It is concluded that in children with prolonged immobilization kidney stone formation may occur with possible significant consequences that should be considered in differential diagnosis. In patients with neurological disease with narrowed logical contact the special attention should be paid for accompanying sparse symptoms.

Authors' contributions/Wkład autorów

MS – essential contribution to the concepts and design work, data collection and interpretation, critical reviewing work for important intellectual content, final acceptance for publication. AZ-B, JM-P – data collection and interpretation. PA, EK – essential contribution to the concepts and design work, critical reviewing work for important intellectual content.

ET-D, ZG – literature search. AP – essential contribution to the concepts and design work.

KZ – critical reviewing work for important intellectual content krytyczne zrecenzowanie pod katem istotnej zawartosci intelektualnej akceptacja ostatecznej wersji do opublikowania, final acceptance for publication.

Conflict of interest/Konflikt interesu

None declared.

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None declared.

Ethics/Etyka

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; Uniform Requirements for manuscripts submitted to Biomedical journals.

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